EXPERT REVIEWS

Measurement of physical activity in patients with cystic fibrosis: a systematic review

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Physical activity (PA) improves exercise capacity, slower decline in lung function and improve quality of life in patients with cystic fibrosis (CF). Despite the importance of PA, it is important to assess the amount of PA. The objective of this literature review was to evaluate the validity and usability of instruments that are used to measure PA in patients with CF. GoogleScholar, ScienceDirect, The Cochrane Library and PUBMED database were searched. All studies that included instruments to measure PA of patients with CF, published from 2000 till June 2012 were reviewed. Eight studies were included in this systematic review. At this moment, there is not sufficient evidence to support incorporation of specific tools to facilitate the PA assessment into clinical practice. Pedometers may offer an inexpensive method of obtaining a measurement of PA, and there is some evidence for supporting its use in CF.

KEYWORDS: cystic fibrosis • physical activity measurements • systematic review

Cystic fibrosis (CF) is an autosomal recessive disorder. It is caused by mutations in the gene-encoding CFTR, which result in reduced chloride secretion and increased sodium absorption. Patients with CF develop respiratory dysfunction in early childhood and have repeated pulmonary infections with viscous secretions, causing airways obstruction, recurrent pulmonary infiltrates and hyperinflation [1–3]. This will lead to a limited exercise capacity and reduced physical activity (PA) levels [4,5], limited by the degree of severity of their lung disease [6]. The respiratory dysfunction often develops in the first year of life and is eventually the major cause of early morbidity and mortality in CF [7]. It appears that over 90% of CF patients decease due to ventilatory failure [7–9]. Tepper and co-workers suggested that the life expectancy of CF patients can be extended if normal lung function is maintained for as long as possible [7]. This is supported by Wilkes and co-workers, who concluded that early treatment and exercise provides advantages to long-term morbidity and survival in children with CF [10]. Patients with a higher exercise capacity have a better prognosis, survival and quality of life [11,12]. PA is independently related to aerobic capacity in patients with CF and other diseases, as well as in healthy individuals [13]. PA contributes to a slower decline in lung function and an improved quality of life in children and adolescent with CF [14]. Children with CF engage in less vigorous physical activities than their healthy non-CF peers, despite having good lung function [15]. In view of the progressive nature of the disease and the association between aerobic fitness and vigorous activity, patients should be encouraged to engage in more vigorous activities that promote aerobic fitness and may ultimately have an impact on survival, because peak oxygen uptake (VO_{2peak}) is related to survival in CF [11].

There are a number of reasons for this, for example, emotional barriers and low lung function [16]. Because of the importance of PA for patients with CF, it is important to assess the amount and intensities of the patients' PAs. To measure PA levels and patterns in the healthy population, accelerometry and questionnaires have been used. The Lipid Research Clinics (LRC), the Seven Day Physical Activity Recall (7D-PAR) and the Habitual Activity Estimation Scale (HAES) questionnaire performed and validated in healthy children and adults and have also been employed in patients with CF [17]. A lot of research has been done on measuring PA; however, it is relatively unknown which instruments are valid and

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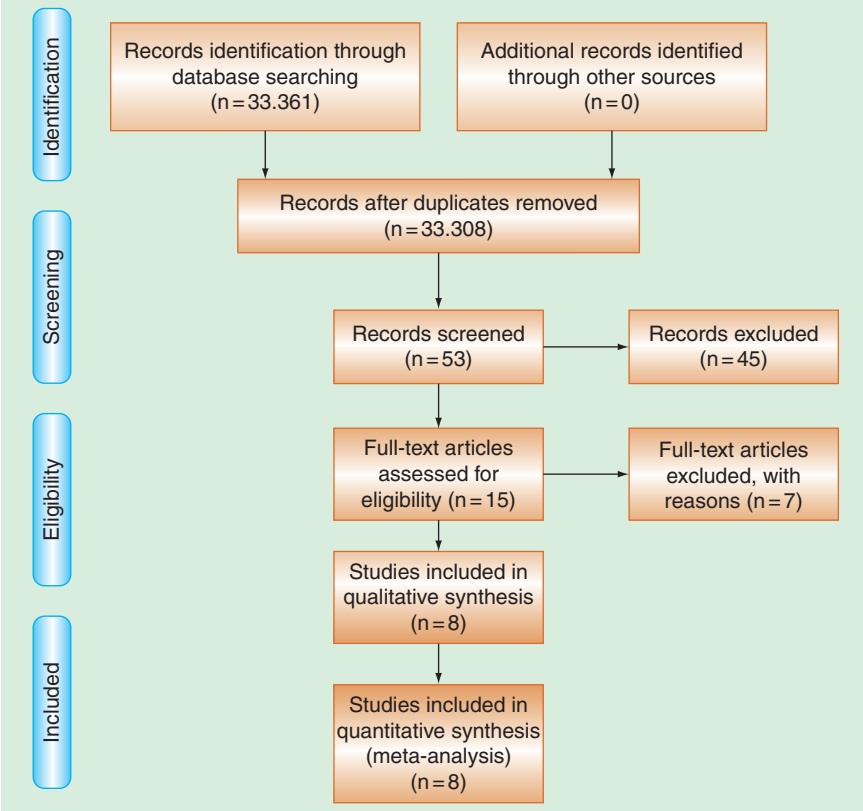

Figure 1. Flow diagram after PRISMA statement.
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usable in patients with CF. Therefore, the objective of this literature review was to evaluate the validity and usability of instruments that are used to measure PA in patients with CF.

Methods

For this review, the PRISMA statement was used, which consists of a 27-item checklist and a 4-phase flow diagram (Figure 1) [18]. A search of all studies, reporting measurement of PA published from 2000 till June 2012 were reviewed using Pubmed, Siencedirect, Cochrane library and Google scholar. Only studies from 2000 were searched because the first study of measuring PA levels in CF was published in 2001 [15].

Studies were identified using the following keyword search terms: CF, asses/measure, (habitual) PA and instruments. The decision to screen articles was based on the article's title and the abstract. Reference lists of all screened articles were scanned to identify studies not evident in the primary literature search. Studies were only included if the study was related to patients with CF and the measurement of their PA. The studies had to be a randomized controlled trial, interventional study, cohort study, case–control study, epidemiologic assessment or survey. Studies were excluded from review if there was insufficient information about the clinimetric quality of the used instrument, and if the study was published before 2000.

Results

Description of the included studies: After searching, 33,361 records were identified, 52 articles were screened and 5 full articles and 3 abstracts met the inclusion criteria and were included in the systematic review. Details of the included articles are presented in Table 1. Two studies were conducted in the UK, two in Australia, USA and in Canada, and one in Spain and Germany. The sample sizes ranged from 9 to 109 patients (age range of 6–42 years) with CF and only one study included a control group. Most studies were performed in children and adolescents and all studies included both genders. Five studies used the HAES, two studies an activity diary, one study a pedometer and five studies used an accelerometer to measure PA. Other questionnaires than the HAES were: the 7D-PAR; the LRC questionnaire; and the International Physical Activity Questionnaire (IPAQ) (Table 1).

Questionnaires

The HAES questionnaire was found as a reliable and valid instrument that can be used to assess activities of varying intensity in patients with CF. The intraclass correlation coefficient (ICC) estimates of reliability for the HAES was 0.72 ($p < 0.001$) [19]. Particularly, time spent in the category 'active' of the HAES was correlated significantly with MVPA (time spent in moderate and vigorous PA) assessed by accelometry ($r = 0.33$) [17]. However, young children found the HAES too difficult to fill out, but it appears feasible for their parents [20]. Adams *et al.* found a poor correlation between the HAES and the Actiwatch ($r = 0.36$) and between the HAES and the peak oxygen uptake ($r = 0.52$) [21]. Schneiderman-Walker *et al.* found a significant correlation between the activity diary and the HAES for total activity in the summer ($r = 0.62$, $p < 0.002$) [14]. They also confirmed that the HAES was a feasible tool for routine follow-up of habitual PA. Ruf *et al.* showed significant correlations between the 7D-PAR and PA assessed by accelerometry in the categories 'hard' and 'very hard' ($0.41 < r < 0.56$) [17]. Also significant ICCs were observed between the 7D-PAR activity categories and MVPA (ICC = 0.40–0.44). Only the LRC questionnaire showed moderate correlations with exercise test parameters [Peak work rate (W_{max}): $r = 0.46$, $p = 0.002$; VO_{2peak}: $r = 0.32$, $p = 0.041$], but the LRC was not related to objectively determined PA. The 7D-PAR may be selected to describe PA within a population. However, the LRC, 7D-PAR and the HAES were not able to generate valid PA data or provide valid assessment of aerobic fitness at the individual level. Because they could

Table 1. Characteristic of reviewed studies.

Study (year)	Instrument	Study population	Results	Conclusion	Ref.
Adams *et al.* (2006)†	HAES Actiwatch (Cambridge Neurotechnology Ltd, UK)	9 CF, 11.3 yr ± 3.3	Mean percentage of awake time spent in moderate or vigorous exercise reported by HAES: 50.9% (±16.3) and measured by Actiwatch: 28.2% (± 9.7). Mean VO_{2peak}: 39.0 (± 7.94) ml kg 1 min Correlations VO_{2peak} and HAES: r 0.52; VO_{2peak} and Actiwatch r 0.32; HAES and Actiwatch r 0.36	Aerobic fitness was moderately correlated with measures of regular activity. Questionnaire and accelerometer measures of activity were correlated but with poor agreement	[21]
Button *et al.* (2010)†	IPAQ Actigraph GTIM	30 CF, 29 yr, 53% fm, FEV1 61%	Spearman correlations of FEV1 % predicted with Actigraph EE: r = 0.68, p < 0.001; and with IPAQ EE r = 0.28, p > 0.05. Correlation of the IPAQ EE with Actigraph EE was moderate (r = 0.46, p = 0.010). There was a trend toward higher EE measured by the IPAQ than measured by the Actigraph (Wilcoxon signed rank test: z = -3.4, p = 0.001). Bland-Altman plot showed poor agreement between energy expenditure from the two measures, with limits of agreement from -11423 to 5550 kcal	The IPAQ underestimates physical activity for patients with lower EE activities and overestimates for those with higher EE in adults with CF. The IPAQ would be a useful clinical screening tool for exercise prescription and monitoring of longitudinal physical activity	[22]
Dwyer *et al.* (2009)	SWA	17 CF, 17 control 26 yr, FEV1 54%	No significant difference in EE measured by IC (6.0 ± 3.4 kcal/min) compared to the SWA estimate (6.3 ± 2.5 kcal/min). The SWA significantly overestimated EE at low exercise intensities and underestimated EE at high exercise intensities. Correlations between EE values, estimated by the SWA and measured by IC, were greater than 0.85 (p < 0.001) for both groups. Standard multiple regression showed that diagnosis of CF independently predicted less than 0.1% of the difference between the IC measure of EE and the SWA estimate. The SWA recorded slightly but significantly fewer steps (113 ± 12 steps/min) than the manual count (119 ± 9 steps/min). r = 0.66, p < 0.001	Diagnosis of CF had no significant negative impact on the accuracy of the SWA estimate of EE. The SWA provided a reasonably accurate estimate of EE and step count during treadmill walking	[23]
Kent *et al.* (2011)†	HAES CFQ-R	16 CF (and their parents), 6–11 yr	All children were unable to complete the HAES, and all able to complete CFQ-R. Mean (SD) score on the CFQ-R: 81.7(12.6) for parent-reported and 80.8(9.2) for child-reported questionnaires. The parent and child CFQ-R were found to be reliable [mean (SD) difference Times 1 to 2: -0.05 (7.5) p = 0.58, 1.84 (6.7) p = 0.29, respectively). No relationship between time spent active and FEV1% predicted (weekdays: r = 2.63, p > 0.05; weekend: r = 2.67, p > 0.05)	Young children found the HAES too difficult however it appears feasible for parents. The CFQ-R and HAES were reliable and there was a trend toward a moderate relationship in parent-reported questionnaires	[20]

†Only abstract available.

7D-PAR: The 7-Day Physical Activity Recall questionnaire; fm: Female; FEV1: % of predicted; HAES: Habitual Activity Estimation Scale; IC: Indirect calometry; ICC: Intraclass correlation coefficient; IPAQ: International Physical Activity Questionnaire; LRC: Lipid Research Clinics questionnaire; SWA: SenseWear activity monitor; yr: Year;

Table 1. Characteristic of reviewed studies (cont.).

Study (year)	Instrument	Study population	Results	Conclusion	Ref.
Quon *et al*. (2012)	Pedometers New-Lifestyles Digi-Walker Pedometer (Model SW-401).	30 CF, 22 yr ± 7	The fair distribution of pedometer recordings throughout the week did not differ by health state (ill versus well: p = 0.76) or age group (12 to ≤18 years versus 19–38 years: p = 0.97). The ICC for daily mean step rate was 0.43 when ill and 0.47 when well. The step rate did not differ significantly day to day within a given 7-day period (ill period p = 0.24; well period p = 0.55). The mean number of hours of use per day was lower during ill compared to well periods, but not statistically significant. Step rate measured by the pedometer was correlated with self-reported physical activity items on the CFRSD	Step rate measured with a pedometer correlates significantly with changes in health and self-reported activity, and could be used as an outcome measure in CF. Pedometers offer several practical advantages over accelerometers as they are easier to use and much more affordable. Pedometer-recorded step rate also correlated with self-reported physical activity items on the CFRSD	[24]
Ruf *et al*. (2012)	HAES 7D-PAR LRC Actigraph (GT1M, Pensacola, FL, USA).	41 CF, 12–42 yr	Time spent in the categories 'hard', 'very hard' and 'hard & very hard' of the 7D-PAR (0.41 < r < 0.56) and 'active' (r = 0.33) of the HAES correlated significantly with MVPA (time spent in moderate and vigorous physical activity). The activity levels of the LRC were not related to objectively determine physical activity. Significant ICCs were only observed between the 7D-PAR activity categories and MVPA (ICC = 0.40–0.44). Only the LRC showed moderate correlations with the exercise test (W_{max}: r = 0.46, p = 0.002; VO_{2peak}: r = 0.32, p = 0.041)	The activity categories 'hard' and 'very hard' of the 7D-PAR best reflected objectively measured MVPA. Since the association was at most moderate, the 7D-PAR maybe selected to describe physical activity within a population. None of the evaluated questionnaires was able to generate valid physical activity at the individual level. Neither did any of the questionnaires provide a valid assessment of aerobic fitness on an individual level	[17]
Schneiderman-Walker *et al*. (2005)	HAES Activity diary	109 CF, 7–17 yr	There was a significant correlation between the AD and HAES for total activity in the summer only (r = 0.62, p < 0.002)	The HAES is a feasible tool for routine follow-up of HPA	[14]
Wells *et al*. (2008)	HAES ActiGraph (TM Tri-Axial) Accelero-meter (model 7164) Activity diary	14 CF, 16.2 yr ± 4.2	ICC estimates of reliability for the HAES, diary, and accelerometer were 0.72 (p < 0.0001), 0.76 (p < 0.0001), 0.63 (p < 0.0001), respectively. Significant relationships between the participants' activity results as estimated by the HAES, diary and accelerometer	The HAES questionnaire is a reliable and valid instrument that can be used to assess activities of varying intensity in patients with CF	[19]

†Only abstract available.
7D-PAR: The 7-Day Physical Activity Recall questionnaire; fm: Female; FEV1: % of predicted; HAES: Habitual Activity Estimation Scale; IC: Indirect calometry; ICC: Intraclass correlation coefficient; IPAQ: International Physical Activity Questionnaire; LRC: Lipid Research Clinics questionnaire; SWA: SenseWear activity monitor; yr: Year;

provide only rough estimates of an individual's objectively measured PA, none of these PA questionnaires were precise enough to be useful for individual counseling [17].

Correlation between the IPAQ energy expenditure (EE) and PA measurement by the Actigraph was moderate (r = 0.46, p = 0.010), but the IPAQ underestimates PA for patients with lower EE activities and overestimates for those with higher EE in adults with CF. There was also poor agreement in EE between the IPAQ and the Actigraph from -11,423 to 5550 kcal [22].

Activity monitors

The activity monitors that were used to measure PA in CF patients were the SenseWear monitor (SWA) [23], the Actiwatch [21], the ActiGraph (GT1M, ActiGraph, Pensacola, FL, USA) [17], the ActiGraphTM Tri-Axial Accelerometer (model 7164) [19] and the New-Lifestyles? Digi-Walker Pedometer (Model SW-401) [24]. Wells *et al.* compared the results of the PA by the ActiGraph Tri-Axial Accelerometer (model 7164) between the first and the second week to determine the reliability and found a ICC of 0.63 ($p < 0.0001$) [19]. Dwyer *et al.* found correlations between EEs values estimated by the SWA and measured by breath by breath indirect calorimetry > 0.85 ($p < 0.001$) [23]. However, the SWA significantly overestimated EE at low exercise intensities and underestimated EE at high exercise intensities. It also recorded a slight but significant lower amount of steps than the manual count. Step rate measured by the New-Lifestyles? Digi-Walker Pedometer (Model SW-401) did not differ significantly between days measured over a 7-day period (ill period p = 0.24; well period p = 0.55) [24]. The mean number of hours of use per day was slightly lower during ill compared to well periods, but the difference was not statistically significant. The moderately pedometer recordings throughout the week did not differ by health state (ill versus well: χ^2 p = 0.76) or age group (12 to ≤18 years versus 19–38 years: χ^2 p = 0.97). Finally, step rate measured by the pedometer was also correlated with self-reported PA items on the CFRSD [24].

Discussion

The objective of this literature review was to evaluate the validity and usability of instruments that are used to measure PA in patients with CF. Different instruments have been used to measure PA and the most commonly used instruments are the HAES and accelerometers. The HAES was found as a reliable and valid instrument, which is in agreement with Hay and Cairney, who concluded the HAES to be an useful clinical tool and for research purposes in adolescents and children with chronic disorders [25]. The HAES cannot be used by every age, but patients >11 years can complete it without assistance, once given proper instruction. It is difficult to fill the HAES out for younger children (<11 years), especially estimating percentage of time spent at each activity level, but it appears feasible for parents.

Furthermore, the HAES is only valid at higher activity levels and cannot accurately measure light PA [14]. Evidence of validity and applications of the HAES is shown by Schneiderman-Walker *et al.* [14]. They used the HAES and showed that declines in lung function among boys with CF were correlated with habitual activity measured by the HAES. Furthermore, the 7D-PAR questionnaire can be used to describe PA within a population and only the LRC questionnaire showed moderate correlations with the exercise test. However, the LRC, 7D-PAR and the HAES were not able to produce valid PA and exercise data at the individual level. Neither did any of the questionnaires give a valid assessment of aerobic fitness on an individual level. However, even the 7D-PAR could make rough estimates of an individual's measured PA. None of the above PA questionnaires were useful for individual counseling. For epidemiological studies, however, a PA questionnaire might be adequate to describe the activities in patients with CF. For individual analysis, additional objective measures such as accelerometry are required.

The IPAQ underestimates PA for patients with lower EE activities and overestimates for those with higher EE in adults with CF. So the IPAQ would only be a useful clinical screening instrument for the longitudinal monitoring of PA. Questionnaires to assess PA are popular in large populations, easy to use and cost-effective. Because the response on questionnaires is subjective, additional objective measures (such as accelerometry) are recommended [26].

There were five different types of activity monitors used, but only three studies investigated the reliability of the instruments. The ICC was high for the Actigraph Tri-Axial Accelerometer (model 7164) and the SWA, and moderate for the New-Lifestyles? Digi-Walker Pedometer. The SWA overestimated EE at low exercise intensities and underestimated EE at high exercise intensities. Further research is needed to investigate the correlation between PA and the ActiGraph Tri-Axial Accelerometer (model 7164). Step rate measured with a pedometer correlates significantly with changes in health and self-reported activity and could be used as an outcome measure in CF.

Pedometers offer several practical advantages over accelerometers as they are easier to use and are much more affordable [24]. The use of pedometers to monitor step counts in adolescents and adults with CF appears to be feasible. In conclusion, the use of pedometers to measure daily step counts in adolescent and adult CF patients is feasible. The recording of daily step rates can be used to detect changes in health status and self-reported symptoms [24]. Pedometer-recorded step rates also correlate well with related items from the CF Respiratory Symptom Diary [27]. The use of pedometers to monitor PA as an endpoint in clinical trials appears promising but warrants further study. Furthermore, pedometers are an inexpensive and simple alternative to activity monitors. However, caution should be applied during slow walking speeds because of undercounting steps [28].

The Dynaport MiniMod, Actigraph GT3X and SenseWear Armband are the most valid monitors to measure standardized physical activities, but are less accurate for slower walking speeds. The Dynaport MiniMod and Actigraph GT3X discriminate best between different walking speeds [27].

This is the first systematic review, which focused on the measurement of PA in patients with CF. PA is very important for patients with CF because it can improve exercise capacity, which contributes to a slower decline in lung function and an improved quality of life. Only eight studies are available about the validity and usability of instruments that measures PA. There is less information about the CFQ-R and the IPAQ because there are only abstracts available for these studies. Details about the Actiwatch that is used by Adams *et al.* and the Actigraph used by Button *et al.* were not available for this reason [21,22].

Clinical implications

Because of the importance of measurement and assessment of PA for patients with CF, clinicians should use instruments to assess the amount and intensities of the patients' physical activities. Based on the most frequently reported outcomes in the literature, a combination of the HAES questionnaire and daily step counts with a pedometer is advised to provide a comprehensive insight in the PA in patients with CF.

Future research

Further research is needed to investigate at what age level the HAES is feasible and at what age parents should be concerned. Which instrument is valid at low activity levels and at an individual level must also be investigated. Thus, in the future, more attention should be directed toward measurements that reflect the objective individual PA levels. Finally, more research is needed to investigate the best combination of objective and subjective measurements for the individual assessment and promoting of PA in patients with CF.

Expert commentary

Regardless of much improvement with PA assessment, limitations concerning the accurate measurement of PA are often increased in young people. These limitations of PA assessment due to changes that occur during natural growth, gender characteristics, as well as a more intermittent pattern of habitual PA in young people. Self-report instruments/questionnaires and movement sensing are currently the most frequently used methods for the assessment of PA in epidemiological research. Habitual EE can be estimated from these measures with varying degree of uncertainty. Nonlinear modeling techniques, using accelerometry in combination with physiological parameters, like heart rate, have the greatest potential for increasing the prediction accuracy of PA EE. Although multisensor systems may be more accurate, this must be balanced against feasibility, a balance that shifts with technological and scientific advances in the future and should be considered at the start of every new study.

Five-year view

Within 5 years, validated PA monitors have become available, in the clinical practice, to measure PA in patients with CF. Furthermore, personalized interventions have become available to stimulate PA in patients with CF, and the impact of these interventions on clinical outcome measures are reported in clinical studies.

Conclusion

Different instruments have been used in the literature to measure PA in patients with CF. At this moment, there is not sufficient evidence to support inclusion of specific tools to smooth the progress the PA assessment in patients with CF into research and clinical practice. Pedometers may offer an inexpensive method of obtaining a measurement of PA and there is some evidence for supporting its use in CF.

Financial & competing interests disclosure

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Key issues

Key points to physical activity assessment

- Questionnaires and diaries are useful to screen physical activity levels in population.
- At this moment, there is not enough evidence to recommend the use of one physical activity questionnaire/diary over another.
- Pedometers may offer a moderately inexpensive method of measuring physical activity, and there is some evidence of supporting its use in cystic fibrosis (CF).

Key points for future research

- To investigate methods of reporting objective physical activity data for categorising categorizing physical activity intensity specifically in CF. To investigate the relationship between physical activity and exercise capacity in CF.
- To investigate the impact of exercise/physical activity interventions on clinical outcome measures.

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